CASE REPORT

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Ancient Schwannoma with perivascular pseudorosettes in a young adult: a case report

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Abstract

Ancient schwannoma is a benign, long standing Schwann cell tumor. It is commonly seen in the middle age and elderly age population, associated with secondary degenerative changes. When these changes are extensive, neuroblastoma and perivascular pseudo-rosettes may be seen. Here we represent a case of a 28 year female with a subcutaneous swelling left elbow. Grossly, single, oval, capsulated tumor measuring (3X1.5) cm received. Cut section showed variegated appearance with areas of hemorrhage and cystic change. Clinicohistopathological features and differential diagnosis revealed a case of ancient schwannoma with perivascular pseudo-rosettes. Due to these unusual morphologic features, a diagnostic confusion has occurred with sarcoma, especially malignant peripheral nerve sheath tumor. So careful observation of the mitotic activity and necrosis is recommended to rule out malignancy.

Key words

Ancient schwannoma, degenerative atypia, histopathology, perivascular pseudo-rosettes, young adult

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Background

Schwannoma or Neurilemmoma is a benign type of tumor originating from nerve sheath. These tumors can originate from any nerve, namely, cranial nerve (except for optic, olfactory), spinal and autonomic nervous system (ANS) which is covered by a Schwann cell sheath [1].

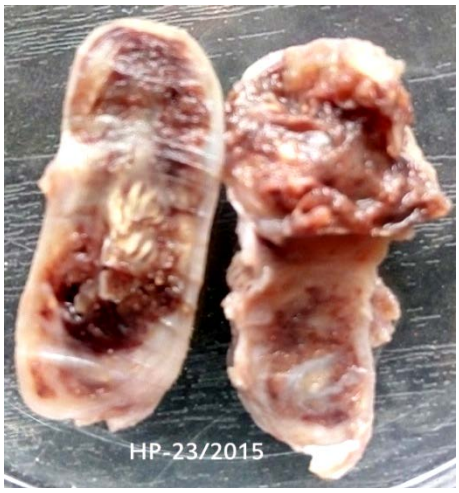
Based on histological structures, schwannomas are categorized into five different types: common, plexiform, cellular, epithelioid and ancient [2]. Ancient schwannoma is one of the several variants of benign schwannoma, which does not show classical antoni A and antoni B areas. "Ancient" refers to demonstrate a group of neural tumours with characteristic pronounced degenerative transformations and diffuse hypocellular area [3]. This is comparatively rare variant of Schwannoma, located deeply as a large mass for a prolonged time and responsible for 0.8% of all soft-tissue tumours [4 - 6].

Distinctive histological degenerative changes are evident in ancient Schwannoma, which is absent in conventional types, such as relative loss of Antoni type A tissue, perivascular hyalinisation, calcification, cystic necrosis, haemorrhage and the presence of degenerative nuclei, sometimes clinically misinterpreted as sarcomatous pleomorphism [7]. These degenerative findings typically occur in long standing lesions, and thus are more commonly found in more deeply situated tumors [6]. These changes are common in benign cutaneous Schwannoma and a considerable factor for vascular abnormalities [8]. In severe conditions these degenerative changes may be associated with neuroblastoma-like and perivascular rosettes. Lewis *et al* in 2005 have noticed only seven cases of schwannomas with giant fibrillar rosettes or perivascular rosettes within a span of ten years [9]. It has also been observed that differentiated Schwann cells immunohistochemically express the S100 protein. So intense immunostaining for S100 suggests a neural origin and a useful diagnostic criteria [5]. Here we report such an unusual variant of Schwannoma with perivascular rosette formation.

Case Report

A 28 year female presented with a subcutaneous swelling left elbow for one year. There was history of trauma two years back. Local examination revealed solitary, soft, movable, nontender swelling measuring (3X1.5) cm. The tumor was surgically removed and sends to the histopathology lab for further investigation. Grossly, single, oval, capsulated tumor measuring (3X1.5)cm received. Cut section revealed variegated appearance with areas of hemorrhage and cystic change (Figure 1). Histology showed a capsulated schwann cell tumor without distinct antoni A and Antoni B zonation, but with focal vague verocay body

Figure 1 - Gross showing capsulated tumor with variegated cut surface



formation (Figure 2A). Degenerative changes like hemorrhage, cystic change stromal hyalinization and thick walled hyalinised blood vessels were seen. Tumor cells showed characteristic features like focal increased cellularity, nuclear atypia and nuclear pseudoinclusion (Figure 2B). The striking feature was perivascular accentuation of pleomorphic tumor cells with formation of perivascular pseudo-rosettes in some places (Figure 2C). Epitheliod appearance of endothelial cells of some of the vessels was seen, but mitosis and necrosis was absent. Based on histopathological characteristics, diagnosis was given as ancient schwannoma with perivascular pseudo-rosettes.

Discussion

The term 'ancient schwannoma' was first introduced by Ackerman and Taylor in the review of 48 neurogenic tumours of the thoracic region. In their report, 10 patients with tumours were presented showing features similar to those of typical neurilemmomas, with some significant differences in tumour portions which contained only a few cells within hyalinised matrices. Authors also noticed that these characteristic changes occurred in schwannomas of long duration [10]. Growth and "aging" of the tumour depends on this degeneration and histologically a vast number microscopic changes like nuclear hyperchromasia, mild nuclear pleomorphism, stromal oedema, hyalinisation and xanthomatous changes apparent in the tissue [11].

Nuclear atypia, pleomorphism & Neurofibromatosis type 2

Despite the degree of nuclear atypia, absence of mitotic activity suggests that it is not malignant nuclear atypia, but a degenerative phenomenon [6].

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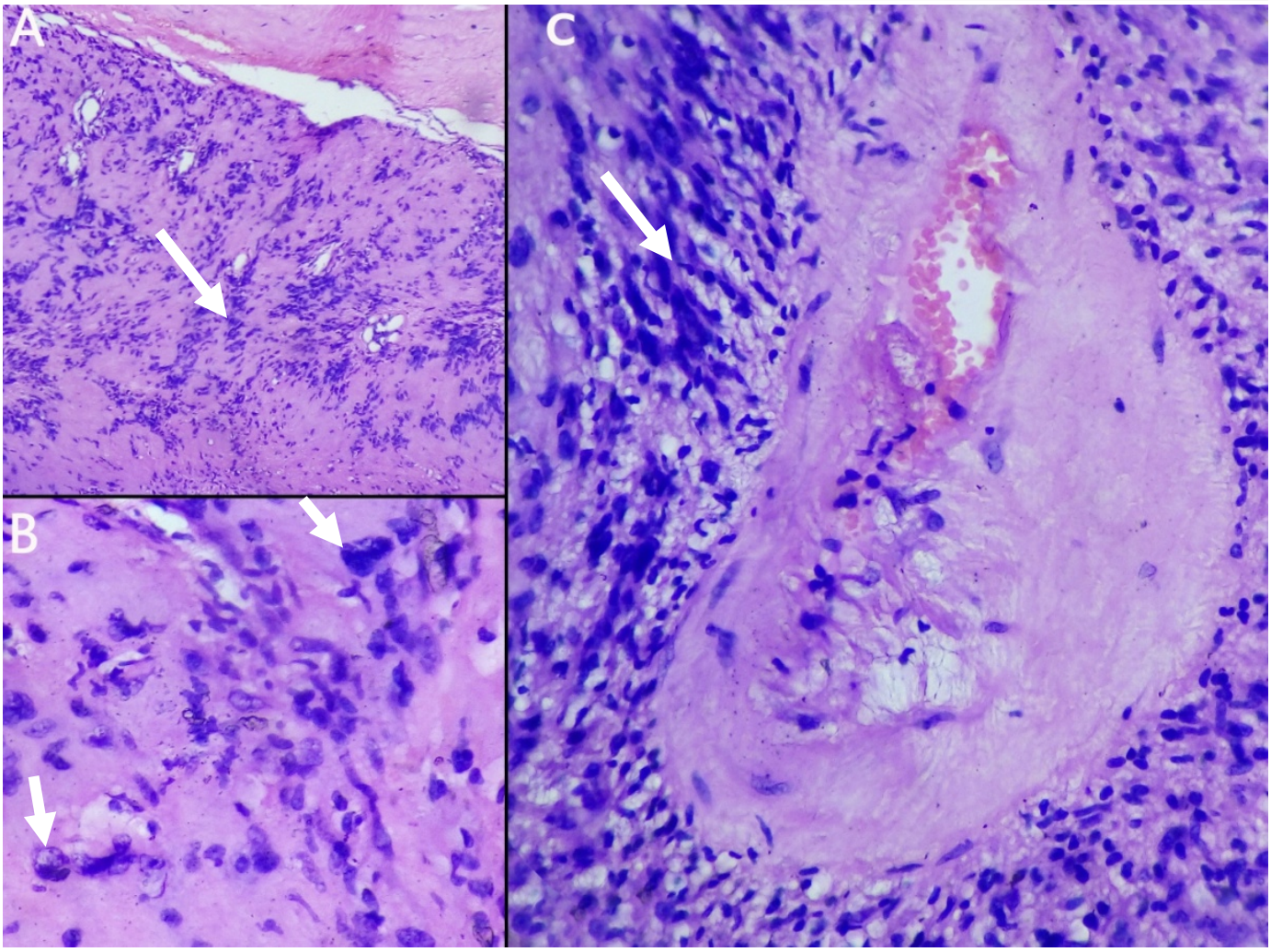

Figure 2 –

A: showing vague verocay body formation (HEX10x)
B: showing degenerative atypia and nuclear pseudoinclusion in tumor cells (HEX40x)
C: showing perivascular pseudo-rosettes (HEX40x)

In our case marked pleomorphism with giant cell formation was seen, but mitotic activity was absent. Approximately 90% of schwannomas are solitary and sporadic, while 4% arise in the setting of Neurofibromatosis type 2 (NF2) and 5% are multiple but unassociated with NF2 [12].
Multiple schwannomas are also seen as a component of schwannomatosis [13]. In the present case the tumor was solitary and there was negative family history for NF2.

Perivascular rosette formation
The most common reported site for schwannoma is skin and subcutaneous tissue as in our case [14]. Schwannoma with rosette formation must be differentiated from neuroblastoma, peripheral neuroectodermal tumors (PNET), and malignant change in a schwannoma i.e. malignant peripheral nerve sheath tumor (MPNST). In this case neuroblastoma and PNET-like rosettes are not seen. The most striking finding was perivascular concentration with rosetting of tumor cells and epitheliod appearance of endothelial cells of these vessels, which raised a suspicion of malignant change *i.e.* MPNST. Although exceedingly rare, schwannomas do have the capacity to undergo malignant transformation; thus created a diagnostic dilemma [15].
When several other features were considered e.g., absence of necrosis, mitosis, and invasiveness, as well as the absence of abrupt transition between typical schwannoma areas and cellular foci of atypical large cells supported the diagnosis of ancient Schwannoma [16].

Conclusion

Ancient schwannoma may show unusual morphologic features like perivascular pseudo-rosettes in addition to its degenerative nuclear atypia causing diagnostic confusion

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with sarcoma especially MPNST. So careful observation is strongly recommended for the mitotic activity and necrosis to rule out malignancy.

Abbreviations

Autonomic nervous system (ANS), malignant peripheral nerve sheath tumor (MPNST), Neurofibromatosis type 2 (NF2), peripheral neuroectodermal tumors (PNET)

Competing interests

None declared.

Authors' contribution

Dr. K M Panda: Concept and design of study interpretation of data, drafting the article or revising it critically for important intellectual content; Dr. Reena Naik: acquisition of data or analysis & interpretation of data, final approval of the version to be published. Dr. PC Agrawal: editing and final approval of the version to be published.

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